Accessory Valvular Tissue Causing Subpulmonary Stenosis in Corrected Transposition of Great Vessels

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The malformation, corrected transposition of the great vessels, has received sufficient attention to make it a relatively simple condition to recognize clinically. Moreover, the clinical manifestations of ventricular septal defect, mitral insufficiency, and pulmonary stenosis—conditions that commonly occur with the basic malformation—are also readily interpreted and have received some degree of coverage in the literature. Little attention, however, has been directed to a peculiar anatomic deformity that may occur in some cases and accounts for obstruction to pulmonary flow.

It is the purpose of this communication to describe, in three cases with corrected transposition of the great vessels, a formation of accessory tissue of the venous atrioventricular valve that was responsible for obstruction to the egress of blood from the venous ventricle in each instance.

The essential anatomic details for the three cases are described first, following which the essential clinical findings and other pertinent observations are presented, including the surgical features of two cases.

Anatomic Findings

The three cases from which the necropsy specimens were obtained are represented by patients aged 7 years (case 1), 4 days (case 2), and 14 years (case 3), respectively, at the time of death. In the first two cases the classical features of corrected transposition of the great vessels in situ solitus were present (fig. 1, left); in case 3 the heart showed the characteristics of corrected transposition of the great vessels in situ inversus.

In case 1 the pulmonary valve was congenitally bicuspid but not stenotic, while in each of the other two cases the pulmonary valve showed three normal cusps.

The anatomic feature of chief interest in each case was located at that region where, in corrected transposition, the venous atrioventricular valve is in continuity with the pulmonary valve. In the two cases with situs solitus this involved structures in the right side of the heart, whereas in case 3 the venous chambers were on the left side.

In cases 1 and 2 a ventricular septal defect was present below the pulmonary valve. In these two cases an umbrella-like formation of accessory valvular tissue protruded into and obstructed the subpulmonary region of the right-sided (venous) ventricle. The tissue was attached by chordae-like strands to pulmonary and atrioventricular valvular tissue and to the anterior and superior edges of the ventricular septal defect. The accessory tissue was so located as to obscure and obstruct the ventricular septal defect, leaving only its posteroinferior aspect open. The effective opening of the ventricular septal defect lay proximal to the obstruction in the ventricular outflow tract (figs. 1, right, and 2).

In case 3 a cusp-like formation of accessory valvular tissue, measuring approximately 2 cm. in diameter with its free edge directed downward, was attached to the ventricular aspect of the anterior leaflet of the venous atrioventricular valve (fig. 3). No chordae were attached to this structure. When pressure was applied to the under aspect of this cusp-like formation, it bulged as a pouch into and obstructed the subpulmonary region of the venous ventricle. In this case, the ventricular septum was intact, while an atrial septal defect at the fossa ovalis was present.

Clinical and Other Pertinent Features of Individual Cases

Case 1

The patient in case 1, a 7-year-old boy, presented as a problem in cyanotic cardiac disease with evidence of pulmonary stenosis and a right-to-left shunt. Physical examination revealed the
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Cardiac apex to lie in the right hemithorax. A grade III-IV (basis of I-IV) harsh pansystolic murmur was heard loudest over the left lower sternal area and was transmitted over the entire precordium. A systolic thrill was maximal at the level of the fourth left intercostal space. A grade II-III high-pitched, blowing diastolic murmur was heard loudest along the upper left sternal border, maximal at the second left intercostal space. The second cardiac sound at the left parasternal area in the second intercostal space was loud and split.

The electrocardiogram showed a prolonged P-R interval for age and rate. The mean manifest electrical axis was of the left deviation type (−75 degrees). The QRS loop in the frontal plane was oriented superiorly and to the left. The mean T vector was −150 degrees, indicating an abnormal discordancy between it and the mean QRS axis. Left ventricular hypertrophy and strain were indicated by the deep S waves (78 mm.) in lead V1, elevation of the ST segment in lead V4, and inversion of T waves with depressed ST segments in the left precordial leads. Additionally, right ventricular hypertrophy was suggested by the relatively tall R wave in lead V1 (fig. 4).

Roentgenologic examination showed the cardiac apex to be on the right. The aortic arch was on the left and the pulmonary vasculature was interpreted as being of normal or less-than-normal prominence. An unusually large prominence was noted above the left cardiac border; this was thought to represent the ascending aorta (fig. 5, left).

When the patient was 5 years of age, selective right ventriculography had been performed in another institution. This had shown corrected transposition of the great vessels with a right-to-left shunt at ventricular level and subpulmonary stenosis. Throughout the cardiac cycle, a persistent zone of narrowing below the origin of the pulmonary trunk was demonstrated (fig. 5, right).

When studied by us 2 years later, conventional cardiac catheterization showed the right ventricular systolic pressure to be equivalent to brachial...
artery systolic pressure but, at that time, the catheter could not be advanced into the pulmonary artery.

At the time of operation, pulmonary stenosis was established with the recording of the following pressures: pulmonary trunk: 25/5 mm. Hg; right ventricle: 170/10-25 mm. Hg; aorta: 150/90 mm. Hg.

With aid of extracorporeal circulation a right-sided ventriculotomy was performed. Through this approach a mass of soft, sponge-like, pale gray accessory tissue was observed to bulge into the outflow tract beneath the pulmonary valve; this partially obscured an existing ventricular septal defect. The exact origin, structure, and function of the accessory tissue remained uncertain and, for that reason, the tissue was not removed. The ventricular septal defect could not be well defined by this approach. Consequently, after the right-sided ventricle was closed, a left-sided ventriculotomy was performed and the ventricular septal defect was closed through that route.

At the same time it was noted that aortic blood was regurgitating into the systemic ventricle through a perforation in one of the aortic cusps. This was overcome by suturing the edges of the perforation.

Twelve hours after an initially favorable postoperative response the patient became hypotensive, developed cardiac standstill, and died.

The necropsy, in addition to showing the obstructive accessory tissue described, revealed a cleft in the anterior leaflet of the venous (right-sided) atrioventricular valve. Regurgitation had probably occurred through this valve, also.

Case 2

The patient, an acutely ill dyspneic female infant, was admitted to the hospital at the age of 3 days.

Pronounced generalized cyanosis was present. The heart was in a normal position. There was a grade II-IV (basis of I-IV) systolic murmur along the left sternal border with maximal intensity at the third intercostal space. The second sound at the "pulmonary area" was loud and split.

Electrocardiograms showed persistent complete atrioventricular block, with a ventricular rate of 40 beats per minute.

Roentgenologic examination revealed cardio-

Figure 2

Case 2. Left. Anterior view of heart, lungs, and great vessels. The ascending aorta (A.), which has been retracted to the right in the preparation of the photograph, lay anterior to the pulmonary trunk (P.). A wide patent ductus arteriosus (D.A.) extends from the pulmonary trunk to the descending aorta. The aortic arch shows tubular hypoplasia between the left subclavian and the left common carotid arteries. Right. Interior of the right-sided (venous) ventricle (V.V.). The probe lies in the ventricular septal defect above which is an accumulation of accessory tissue (A.c.); this attaches both to the right atrioventricular valve (A.V.) and adjacent pulmonary valve (P.V.).
megaly and a minimal increase in the pulmonary vascular markings (fig. 6). A peripheral venous forward angiocardiogram showed simultaneous filling of transposed great arterial vessels. The pulmonary trunk lay posteriorly and was relatively wide. The ascending aorta was well opacified, and was observed to arise anteriorly. Opaque medium then appeared in the descending aorta but the arch remained unopacified. No abnormalities were revealed at the semilunar valvular or subvalvular levels by this study.

On the basis of the findings it was considered that an intracardiac right-to-left shunt, associated with some form of transposition of the great vessels, represented the essential problem. Coarctation of the aorta or interruption of the arch was also suspected.

The patient did not respond to intensive medical therapy. Her condition remained critical and, on the day following admission, the infant died.

In addition to the crucial anatomic findings already given, this patient exhibited a patent ductus arteriosus and hypoplasia of the aortic arch between the left common carotid and subclavian arteries.

Case 3

This subject was a 14-year-old acyanotic girl who was known to have total situs inversus and a cardiac murmur.

The cardiac apex was located to the right of the sternum. A thrill was felt at the right sternal border and a grade-IV (basis of I-IV) harsh systolic murmur was present over the entire precordium with maximal intensity at the upper right sternal border. Also, a grade-II systolic murmur was located at the apical area (right) and was transmitted to the right axillary region.

The electrocardiogram showed signs of mirror-image dextrocardia and probable biventricular hypertrophy (fig. 7).

Conventional radiologic examination of the thorax revealed the cardiac apex to be on the right side. A prominent shadow along the right upper lateral cardiac border was interpreted either as that of the pulmonary trunk or of the ascending aorta—in the expected position for the aorta in corrected transposition in situs inversus (fig. 8).

Cardiac catheterization indicated a "left-to-right" (arteriovenous) shunt at the atrial level. In the venous ventricular chamber, which lay pos-
teriorly and toward the left, the pressure was 103 mm. Hg systolic and 0 diastolic. The oxygen content of the blood in this chamber was equal to that in the venous atrium. The catheter could not be advanced across the pulmonary valve. Opaque medium was injected into the venous ventricle following which opacification of the ventricle and of the pulmonary artery occurred.

On initial study it was thought that no pulmonary stenosis was present, but restudy of the films revealed that a filling defect was present in the subpulmonary area of the venous (left-sided) ventricle (fig. 9). Catheterization of the arterial ventricle was then performed in a retrograde manner. Instillation of radiopaque medium into this ventricle was followed by opacification of the aorta and the arterial atrium. The pattern outlined was that of corrected transposition in situs inversus and insufficiency of the systemic (arterial, right-sided) atrioventricular valve. Neither intraventricular study showed evidence for a ventricular septal defect.

With a diagnosis of corrected transposition of the great vessels in situs inversus, atrial septal defect, and possible pulmonary stenosis, the patient was operated upon by the use of cardiopulmonary bypass. Through the venous atrium, a large (3.5 cm.) atrial septal defect at the fossa ovalis was found and closed. The arterial atrioventricular valve was found to be slightly incompetent. Next, a pulmonary arteriotomy was performed and the pulmonary valve was observed to be normal. With the experience of case 1 as background, accessory valvular tissue in the subpulmonary region was sought as a possible cause of subpulmonary stenosis but none was identified. While some septal tissue in the wall of the venous ventricle was excised, a demonstrated pressure differential between the venous ventricle and the pulmonary trunk remained essentially unaffected.

Immediately following the operation, the patient developed pulmonary edema which could not be overcome. Death occurred on the fifth postoperative day.

In addition to the features of corrected transposition in situs inversus and the accessory tissue causing subpulmonary stenosis (features that have been described), the necropsy revealed a cleft between the septal and the anterior leaflets of the arterial (right-sided) atrioventricular valve. This was considered to be the basis for the incompetence of this valve which had been observed at operation.

Discussion

The anatomy of congenital corrected transposition is well known. Associated anomalies, when present, dictate the altered hemodynamics and the requirements of surgical therapy.

Associated anomalies include ventricular septal defect, deformities of the arterial atrioventricular valve (causing insufficiency of this valve), pulmonary stenosis, patent ductus arteriosus, and interference with atrioventricular conduction. In corrected transposition when obstruction to pulmonary arterial flow occurs, the anatomic causes previously recognized include pulmonary valvular stenosis or atresia, or subpulmonary muscular or membranous stenosis.

In each of the three cases of corrected transposition here reported, accessory tissue attached to the venous atrioventricular valve was responsible for subpulmonary stenosis.
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We are not aware of an existing description in the literature of this phenomenon.

In our first two cases, each having a ventricular septal defect, the obstructive tissue was situated predominantly above the ventricular septal defect. As a consequence of the subpulmonary stenosis, a right-to-left shunt through the ventricular septal defect is considered to have occurred in each of these two cases. Without the obstruction, it would have been expected that the shunt through the ventricular septal defect would have been in a left-to-right direction.

In the third case, an example of corrected transposition in total situs inversus, an atrial septal defect was associated with an intact ventricular septum. Right ventricular hypertension was recorded which, ultimately, was explained by the obstructive effect of the accessory tissue described.

It is apparent from these specimens that the accessory valvular tissue was potentially resectable. In the second case no operation was performed; hence, this problem had not been met by the practical test. In the first case it was apparent at operation, while the venous ventricle was open, that an obstructive lesion existed in the subpulmonary area. The exact nature of this, however, was not recognized and, consequently, this tissue was not removed. In retrospect, removal of it appears to have been the indicated procedure for relief of the subpulmonary stenosis.

In the third patient it is of interest that during the operation a basis for subpulmonary stenosis was specifically sought through the pulmonary arterial approach. Yet no such lesion could be identified. The probable basis for this oversight is the fact that, when the ventricle was inspected while empty, the accessory tissue (having a flap-like structure) lay flattened against the atroventricular leaflet from which it arose.

It is presumed that, during systole in the intact beating heart, this tissue ballooned and caused subpulmonic stenosis. In this patient

**Figure 5**

Case 1. Left, Posteroanterior roentgenogram of thorax. Transverse size of heart within normal limits. Unusual configuration of the heart showing dextroversion and prominence of the ascending aorta. The latter forms the convexity above the left cardiac border. Right. Anteroposterior view of selective right ventriculogram. Tip of the catheter is in the right-sided (venous) ventricle (VV). Arrow points to the site of subpulmonary stenosis. Pulmonary valve (PV) is well outlined. Contrast medium from the venous ventricle is seen streaming across the ventricular septal defect (VSD) into the left-sided (arterial) ventricle. The pulmonary trunk (PT) occupies a medial position. (Film supplied through the courtesy of Dr. F. A. Hernandez.)
(case 3), clearly, death appeared to result from acute pulmonary edema. This circumstance emphasizes the fact that even mild "mitral" insufficiency may be lethal when allowed to exist after a defect at the atrial level has been closed. Closure of an atrial septal defect usually raises the left atrial pressure; ordinarily, however, this does not rise to a level sufficiently high as to cause pulmonary edema. Under circumstances when mitral insufficiency does exist, as in this instance, the pressure may readily rise to levels precipitating pulmonary edema.

It is recognized that in corrected transposition, the interrelationships between the semilunar and atrioventricular valves and the ventricular outflow tract on the venous side bear considerable structural similarity (in mirror image) to comparable structures of the arterial side of normal hearts. For this reason it is pertinent to mention subaortic obstruction caused by anomalous mitral tissue in normally oriented hearts.

In a case of congenital left ventricular-right atrial communication, Ferencz observed that anomalous chordae tendineae extended from the mitral valve and attached to the septal wall of the left ventricular outflow tract, causing subaortic stenosis. Björk and his co-workers reported on two patients in whom subaortic stenosis was caused by mitral valvular tissue that was deformed and displaced toward the left ventricular outflow tract.

In a case of Lauer and associates, a ventricular septal defect was associated with a cleft in the anterior leaflet of the mitral valve. The anterior half of this leaflet was so fixed by chordae to the ventricular septum as to cause obstruction to left ventricular outflow.

A phenomenon more closely allied to the accessory valvular tissue found in the venous ventricles in our cases is that recently observed by MacLean and associates. In their case with a normally oriented heart, subaortic stenosis was caused by an umbrella-like for-
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The obstruction lesion in the left ventricular outflow tract of the normally oriented heart in MacLean's case is an exact counterpart of the subpulmonary stenosis in our specimens with corrected transposition. In this same case, resection of the accessory tissue resulted in elimination of the subaortic stenosis while competence of the mitral valve was maintained. The necropsy specimen of a case in which subaortic stenosis was caused by an anomaly like that in MacLean's case recently has been submitted by Dr. C. P. Deal, Jr., to the Cardiovascular Registry of The Charles T. Miller Hospital.

It should be mentioned that in the normally oriented heart, accessory tissue of the tricuspid valve also may be present. Neufeld and his group described three cases of the tetralogy of Fallot in which accessory tricuspid tissue had obstructed the ventricular septal defect and led to dynamics that simulated the presence of a small ventricular septal defect in association with pulmonary stenosis.

Pulmonary stenosis in hearts with corrected transposition tends to be caused by more complicated anatomic arrangements than does pulmonary stenosis in the normally oriented heart. In the latter, when the ventricular septum is intact, the obstruction is usually within the pulmonary valve. When a defect is present, the pulmonary stenosis is usually part of the tetralogy of Fallot. In this condition, there has been ample description of the infundibular and valvular stenosis that occurs.

Contrariwise, in hearts with corrected transposition, pulmonary stenosis may be entirely at valve level although this phenomenon appears to be uncommon. In our experience it is more common to find that hearts showing corrected transposition and pulmonary stenosis (whether or not the ventricular septum is intact) will manifest some form of subpulmonary stenosis. The obstruction may simply take the form of a deformity of the muscular part of the ventricular septum—a condition comparable to simple subaortic stenosis of normally oriented hearts.

At other times, as in the three cases herein described, accessory valvular tissue may cause the obstruction. The location of this obstructive lesion appears best to be identified by selective angiography made from the venous ventricle.

In case 2, a forward angiogram was made. The study showed the presence of corrected transposition of the great vessels, a ventricular septal defect, and a patent ductus arteriosus. It was noted on the study that the ascending thoracic aorta filled through a wide patent ductus arteriosus immediately following opacification of the pulmonary trunk. Because of the nature of the study, the contrast medium in the right atrium and right atrial appendage obscured the details of the outflow portion of the venous ventricle, hence the presence of the subpulmonary stenosis was not recognized. This frequently occurs in forward angiography and the advisability of obtaining selective studies from ventricular chambers whenever possible is to be emphasized.
Summary

This study reveals three cases of corrected transposition (one with total situs inversus) where an accessory flap or umbrella-like formation of the venous atrioventricular valve presented into the outflow tract of the venous ventricle causing severe subpulmonary stenosis.

In one case the ventricular septum was intact, while in each of the other two, a ventricular septal defect lay below the obstruction. The latter combination effected a right-to-left shunt at ventricular level.

These cases are important because they portray anatomic details of a condition which, although uncommon, is potentially curable.

Clinical identification of the subpulmonary stenosis appears best to be accomplished by selective angiocardiography at the level of the venous ventricle.

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